

13.

Malformation of arm.

By ALLEN STURGE, M.D.

HERBERT A—, æt. 22, admitted as a patient at the Royal Free Hospital for some trifling ailment, presented a congenital deformity of the left arm, belonging to the class of malformations which have generally been described as due to intra-uterine amputation.

Family history.—His father, who was a healthy man, died at the age of 71. His mother living and healthy. She asserts that when pregnant with the patient, she was frightened by a man who had lost an arm, and who ran after her in a wood. The patient has a brother and three sisters, all of whom are healthy and present no deformity of any kind. A brother, a soldier, died of "tumour of the brain." Two other children died in infancy.

The patient is a very fine man, his height being 6 ft. 3½ in., and he is well made in all respects except the deformity of the arm. The greater part of the left forearm and the hand are absent, the forearm ending in a somewhat conical stump about three inches below the elbow-joint. Both the radius and the ulna are present in this stump, the upper extremities of these bones being apparently normal. The lower end of the bones, which apparently do not articulate with one another, taper off into points, the tapering extremity of the radius being rather longer and more sharply pointed than that of the ulna. Over the end of the radius the skin forms a conical projection, the size of a hazel nut, which stands out prominently from the general surface of the stump. There is no adhesion between the skin and the bone. The skin over the end of the ulna is umbilicated, the result, apparently, of adhesion of the subcutaneous tissue to the end of the bone. The skin over the end of the radius presents two faint whitish lines at right angles to one another, one of them being about half an inch and the other about a quarter of an inch in length. These appear to be old cicatrices. There are other cicatrices on the stump, which have the appearance of having resulted

from large superficial sores. The patient says they are the remains of boils, several of which he has had on the stump.

On the flexor aspect of the stump, about midway between the ends of the radius and ulna, are five little projections, situated transversely across the stump. The projection nearest to the radial side of the forearm is the largest, and the others get smaller by degrees as the ulnar side is approached. The largest is about the size of a pea, and the smallest that of a moderate-sized pin's head. The largest and the adjoining nodules have well-marked nails, the free convex surface of which is directed towards the extensor aspect of the forearm. The nail on the second nodule is larger than that on the largest nodule. There is a semblance of horny growth on the third nodule also.

There is free movement of the stump. He can flex the elbow to an angle of about 60° , and he can extend it in an unusual manner, for when the elbow is fully extended the stump of the forearm, instead of being in a straight line with the arm, makes an angle with it posteriorly of about 120° . He seems to have very little power of pronation and supination independently of flexion and extension, but flexion is accompanied by a certain degree of pronation, and *vice versa*. Pronation and supination of the bones can be easily produced by external manipulation.

The muscles of the upper arm and shoulder are considerably smaller on the left than on the right side, and there is shortening of the humerus, to the extent of about half an inch, as compared with the right.

Remarks.—This case comes within the category of those which have been described by Montgomery, Simpson, Annandale, and others, as resulting from intra-uterine amputation. The amputation is attributed to the constriction exercised on the limb by bands of organised lymph due to inflammation in its neighbourhood. These bands having surrounded the limb in an early stage of intra-uterine development, gradually became tightened as the lymph contracts and as the limb grows larger, until at last they bring about a separation of that portion of the member below the seat of constriction.

That this is the true explanation of a certain number of deformities of this kind is proved by the fact that in some few instances the amputated part has been actually found; but I think that a reference to a number of cases of congenital deficiency of parts of limbs will

show that in a considerable proportion of such cases the cause must be sought in some other direction than the hypothesis of amputation *in utero*.

This other mode of causation may consist of either one of two factors—(1) a primary inherent abnormal condition of the ovum, whereby its healthy development is interfered with in one or more directions; (2) a localised morbid condition of one or more parts of the embryo due to onset of disease in the embryo at some period after conception. The former of these two conditions must be invoked in explanation of many cases of monstrosity, of cases where there is congenital hypertrophy of parts, and it is the most probable explanation of supernumerary parts. There can be no reason, therefore, why it should not also produce the opposite conditions, viz. congenital atrophy of limbs and congenital absence of portions of limbs. On the other hand, an inflammatory process capable of throwing out organised lymph in sufficient quantity to produce intra-uterine amputation is quite as likely, if it attacks that portion of the blastoderm from which a limb is developed, to kill the delicate embryonic cells, or to modify their nutrition so profoundly that their subsequent development will be gravely compromised.

It is conceivable, therefore, that cases like that I have described may be produced by mal-development due to either of these two causes.

Mr. Annandale, in his work on 'Malformations, Diseases, and Injuries of the Fingers and Toes,' mentions thirteen cases of congenital deformity, characterised by absence or atrophy of portions of limbs. Four of these he attributed to intra-uterine amputation. He makes no reference to the causation of the remainder, but he would probably attribute them to one of the two above-mentioned causes.

Of the four cases attributed to amputation two were marked by absence of one hand, the arm stopping short at the wrist. In both instances nodules were present on the stump, representing the digits. In a third case absence of the forearm and the occurrence on the stump of nodules representing the fingers was associated with very marked atrophy of the rest of the arm, which was scarcely larger than that of a new-born child. In the fourth case two of the fingers of the left hand were absent, and a third was constricted at the base. In this case all the other limbs presented deformities of some other kind.

Among the cases not referred to amputation was one of absence of both forearms and hands, each stump having one finger growing from it by a loose attachment. There were three cases of absence of several fingers of one hand, two cases of absence of fingers or parts of fingers in both hands, one of absence of fingers in one hand combined with atrophy of the whole arm, one case of absence of certain fingers in both hands, distortion of such fingers as were present, and malformation of one foot.

If these two classes of cases be carefully compared it will be found that there is no feature in connection with the first-mentioned class which justifies us in separating the cases of which it is composed from those forming the second class. For instance, the case of so-called amputation of the forearm in the first class very closely resembled the case in the second series, where both forearms were absent, the only essential differences being that the lesion was bilateral in the latter case, and that in the former case the whole limb was deformed, this latter character pointing rather in the direction of modified development than of intra-uterine amputation. The two instances in which one hand was absent differed also from the case of bilateral absence of the forearm in respect of the unilateral nature of the lesion alone. In the fourth case, attributed to amputation, there is nothing in the description to separate it from others of the same kind placed by Mr. Annandale in the second class; whilst the association of deficiency of certain fingers in one hand, with deformities of other kinds in all the other limbs, argues strongly in favour of mal-development as the cause of all the abnormal conditions present. In two of the four cases referred to amputation, therefore, there is internal evidence in the cases themselves that they were due to mal-development rather than to amputation; whilst the other two instances differ only from cases not classed with the amputations by being unilateral instead of bilateral. The question arises, therefore, whether we ought not, in the vast majority of instances, to look for the cause of congenital deficiency of parts of limbs in the direction of mal-development of the embryo, and whether we ought not to consider intra-uterine amputation as an altogether exceptional cause of such conditions. In certain instances the latter mode of causation can be excluded—those cases, namely, in which absence of a part of one limb is associated with deformity of other limbs, or with great atrophy or other deformity of such parts of the limb as are present. Where a part of the limb is absent, and

the rest of the body, including the upper part of the limb itself, is healthy, the diagnosis of the mode of causation is more difficult; but a comparison of a large number of cases of congenital deficiency will show that every variety may be met with, from cases in which all the limbs are deficient or deformed in some other way, to cases in which the extremity of one finger only is deficient. As, therefore, no line of demarcation can be drawn, dividing the cases into two well-defined groups, so it will, I believe, be impossible to attribute those cases lying at one end of the scale to a cause essentially different from that to which the cases at the other end of the scale have been attributed.

There are two factors which may possibly be found to help us in coming to a correct conclusion with regard to certain of the doubtful cases, and to which, therefore, I think, more attention should be paid. These are—(1) the presence of rudimentary fingers on the end of the stump; (2) the presence of umbilications over the ends of the bones.

I think it is worthy of inquiry whether rudimentary fingers have been present in any of those cases in which the amputated limb has been found. It is very easy to account for them on the hypothesis of mal-development, for in that case they represent the amount of vitality left in the embryonic cells, from which the extremity of the limb should have been developed. On this hypothesis we should expect to find, as, in fact, have been found, many degrees of development, ranging from minute nodules representing fingers at one end of the scale, up to extremities of limbs, which differ but little from the hand, at its other end. This last condition was present in one of the cases recorded by Mr. Annandale. If it be found that no rudimentary fingers occur in undoubted cases of amputation, then their presence would afford presumptive evidence in favour of the hypothesis of mal-development.

As regards the question of umbilication of the skin over the ends of the bone, with which may be classed the presence of scars on the stump, the question arises, "Do umbilication and scarring necessarily indicate amputation?" This question can only be answered by a careful examination of cases in which multiple deformity is present, so as to exclude the hypothesis of amputation. There is certainly no *à priori* reason why the formation of a rounded stump, after destruction of the embryonic cells from which the extremity of a limb should be developed, should not give rise to the appearances of cic-

trisation any more than the healing of a stump after amputation. The skin in such a case would have no necessary tendency to develop uniformly round the end of the limb, and in order for it to cover in the stump of the bones and muscles some unusual process will have to be introduced, which may very possibly give rise to umbilication or slight scarring.

Further attention to these two points is, I think, called for.

The presence of deep depressions round a finger or a limb need not necessarily imply constriction by a band, for mal-development shows itself in curiously partial and localised ways, and there is no reason why it should not be present over a small area in the centre of a limb, in the same way that it is present over a small area at the tip of one finger. If there be disease of the embryonic cells, which should develop into the proximal phalanx of a finger, and not of those from which the two distal phalanges should develop, the proximal phalanx, or part of the phalanx, will be very small, whilst the other phalanges may be of full size. *May 4th, 1880.*